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Hearing loss among children with sickle cell anaemia in Uganda

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ABSTRACT

Background: A case control was carried out to study the prevalence, the pattern and associated factors of hearing loss among Ugandan children with sickle cell anaemia.

Methods: One hundred and thirty-two children known sickle cell and one hundred thirty aged matched non-SCA controls were recruited after medical ethics committee approval and informed consent. Two obtain information on socio-demographic characteristics, history, and examination of the participants, a structured questionnaire was administered. Pure tone audiometry (PTA) was used to assess the hearing in a sound-treated room.

Results: Twenty-two of the children had hearing impairment compared to 6 controls. Prevalence of hearing loss found among children with SCA was 17% but in the control group the prevalence found was 5%. Sensorineural hearing loss (SNHL) affected 39%, conductive (CHL) 25% and mixed (MHL) 14% among cases compared to CHL 7%, SNHL 7% and MHL 7% among the controls. The hearing loss varied from mild to moderate (95%). All sickle cell children had high-frequency hearing loss. There was a statistically significant association of hearing loss and neurologic motor deficit.

Conclusions: Sickle patients are at risk of developing hearing loss as the study demonstrated a difference in hearing threshold in children living with sickle cell anaemia and the controls. High frequencies were more affected. The neurologic motor deficit was highly associated with hearing loss among patients with SCA compared to controls.

Keywords: Sickle cell disease, Hearing loss, Children

INTRODUCTION

Hearing impairment is the loss of the ability to perceive sound in the normal range of hearing.

World Health Organization (WHO) defines hearing disability for children (0–14 years of age) as hearing loss greater than 30 dB in the better hearing ear. ¹

Sickle cell disease is one of the well-documented causes of hearing impairment.² Sickle cell disease is a genetic disorder characterized by the presence of a mutated form of hemoglobin known as hemoglobin S. This abnormal haemoglobin leads to a series of changes which ultimately result in chronic haemolytic anaemia, a

chronic inflammatory state and vasculopathy in both large and small blood vessels.³

Literature has demonstrated a prevalence rate of 11-41% for sensorineural hearing loss (SNHL) in children and adult sickle cell patients. The prevalence of hearing impairment reported in the literature varies to losses of mild to profound hearing levels. Studies done in 2004 in Nigeria revealed the prevalence of 13.4% of sensorineural hearing loss among children with sickle cell disease while in 1996 in Kenya it was found to be 40%. Most of the cases have bilateral hearing loss. The mechanism is not well understood but it seems to occur as a consequence of the cochlear high sensitivity to vessel occlusion which characterizes this disease. In fact, the

sickle cells prevent blood flow to the epithelium of the cochlea by causing ischemia to the stria vascularis. The labyrinthine artery constitutes the one single arterial main blood supply of the cochlear; this makes the inner ear vulnerable to circulatory changes. Studies have reported that the deformation of the red blood cell causes the lesion to the cochlear. This prevents proper blood supply to the metabolic activity required to maintain the ionic and electrical balance of the endolymph. So the anoxia caused to the organ of Corti would cause progressive and extensive damage to the cochlea.

Sickle cell anaemia is still a public health issue in Uganda. In one survey conducted in 2014, it was found that the overall prevalence of sickle cell trait among babies is 13.2% with geographical differences. The south-western region of Uganda had the lowest prevalence (4.6%), while the East central and the Mid North had the highest (19.8% and 19.2% successively). 12 Although the prevalence of sickle cell is high in Uganda, little is known about the prevalence and pattern of hearing impairment among Ugandan children with sickle cell anaemia. The aim of this study was to determine the prevalence, the pattern and associated factors of the hearing loss in children with sickle cell disease. The result of this study provided information about the burden of hearing loss among children with sickle cell anaemia and constituted an evidence-base for clinicians to assess the hearing of patients with sickle cell anaemia.

METHODS

This was a case-control study, where sample size determination was done using Fleiss formula. The study was conducted from May to June 2017. A hearing assessment of 132 sickle cell patients who were systematically recruited from Mulago sickle cell clinic was done. The clinic on average treats 200 patients a week. A total of 67 patients were males (51%) and 64 were female (49%) with age ranging from 5 to 12 years. An age-matched control group of 132 children without sickle cell disease were recruited from general clinics of the Department of Pediatrics of Mulago National Referral Hospital (MNRH). This hospital is affiliated with Makerere University as a teaching hospital for the College of Health Sciences. It has a bed capacity of 1500 beds. The hospital is located in Kampala, the capital city of Uganda. It provides diagnostic, curative, rehabilitation, and preventive services for the whole country and neighboring countries.

Selection criteria

Inclusion criteria

Cases includes sickle cell patients aged 5 to 12 years attending Mulago sickle sell slinic, children whose care takers consented if below 8 years, children who assented and caretakers consented if 8 years and above.

Controls includes non sickle cell patients aged 5-12 years attending Mulago paediatric clinics, children whose care takers consented if below 8 years, children who assented and caretakers consented if 8 years and above.

Exclusion criteria for cases and controls

Patients with history of TB, HIV, meningitis, mumps, ear discharge and history of ear surgery, diabetes, head injury, head and neck radiotherapy, intake of ototoxic drugs and noise exposure. Patients from family with hearing loss, patients who were very sick: patient acutely ill (with acute stroke, acute pain etc.).

Examination to obtain information on socio-demographic characteristics and history of items related to hearing loss and associated factors of the participants; a semistructured questionnaire was administered to the cases and the controls. The otolaryngology examination was carried out by the first author. The pure tone audiometry (PTA) was done by an audiometrician in a sound isolated audiometric room (booth) with ambient sound less than 35 dB using a calibrated diagnostic audiometer KAMPLEX KLD 21 at frequencies 250 Hz, 500 Hz, 1 kHz, 2, 3, 6 and 8 kHz. Hearing loss was defined as present if a child has an average threshold of hearing more than 25 dB at two or more frequencies by PTA in one ear. Data collected was entered using the epidata 3.1 statistical package. Stata version 13.0 statistical software (College Station, Texas).

RESULTS

The audiograms were analyzed into conductive, sensorineural and mixed hearing loss. The hearing was considered normal when the hearing threshold was <25 dB and impaired if >25 dB. The degree of hearing loss was classified as per WHO guidelines (2008): mild degree when the average threshold of hearing is between 26 to 40 dB, moderate if 41 to 60 dB, severe if 61 to 80 dB and profound if more than 81 dB at frequencies 0.5, 1, 2 and 4 kHz. Configuration was determined from the pattern of hearing loss across frequencies. Those in frequencies (0.5, 1, 2 kHz) were regarded as low-frequency losses while those in frequencies (3, 4, 6, 8 kHz) as high frequencies.

Demographic characteristics of 264 cases and controls attending Mulago National Referral Hospital (MNRH)

Age

264 participants were enrolled in the study aged 5 to 12 years with 132 as cases and 132 as controls. The mean age for the cases and the controls was 8.3±2.3 years. Majority of the SCA patients and the controls who participated in the study were between the ages 5-10 years 105 (80%). They were followed by those who were 11-12 years 27 (20%).

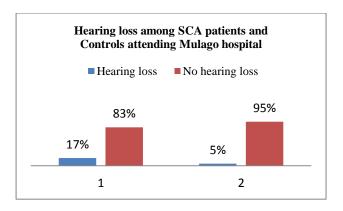


Figure 1: Hearing loss among cases and controls attending Mulago National Referral Hospital (MNRH).

Table 1: 28 participants of cases and controls of hearing loss.

	Cases (n=28) N (%)	Controls (n=28) N (%)
Hearing loss to both ears	19 (86)	5 (83)
Hearing loss to right ear only	1 (5)	0 (0)
Hearing loss to left ear only	2 (9)	1 (17)

Table 2: Distribution of degree of hearing loss.

Hearing level (dB)	Case group (N=28)		Control group (N=28)		
	Right ear	Left ear	Right ear	Left ear	
	Count (%)	Count (%)	Count (%)	Count (%)	
Mild HL 26-40	5 (18)	10 (45)	2 (7)	3 (11)	
Moderate HL 41-60	3 (11)	3 (14)	0 (0)	1 (4)	
Severe HL 61-80	0 (0)	1 (4)	0 (0)	0 (0)	
Profound ≥81		0 (0)	0 (0)	0 (0)	

Sex

In our study 67 patients were males (49%) and 64 were females among the cases which represent successively 51% and 49% of the participants; among the controls, 69 participants were males and 63 were females which are respectively 52% and 48%.

Prevalence of hearing loss

The hearing loss prevalence among cases is 17%, compared to 5% of hearing loss prevalence among controls at a 95% confidence interval.

Cases and controls with hearing loss

Nineteen SCD patients had hearing loss in both ears which means 38 ears, one in the left and two in the right compared to 5 controls who had hearing loss in both ears (10 ears in total), one in the left and none in the right. There were 41 ears with hearing loss among the cases and 11 ears affected among the controls. However, the difference in the side of affected ears between cases and controls is not statistically significant. From the reviewed literature, there is no reported relationship between the side of the affected ear and hearing loss among SCA patients.

Type of hearing loss among cases and controls attending Mulago hospital

The total number of participants (n=28), with hearing loss among them 22 cases and 6 controls.

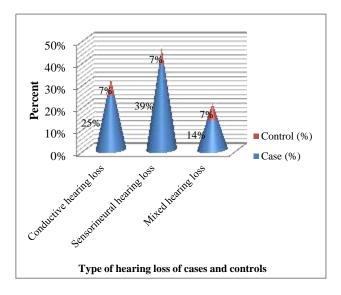


Figure 2: Distribution of types of hearing loss.

Table 3: Distribution of configuration of hearing loss.

Frequency	Cases (n=28)	Controls (n=28)
	Count (%)	Count (%)
500	0 (0)	0 (0)
1000	0 (0)	0 (0)
2000	0 (0)	0 (0)
3000	4 (14)	0 (0)
4000	2 (7)	2 (7)
6000	5 (18)	3 (11)
8000	11 (39)	1 (4)

The degree of hearing loss among cases and controls attending Mulago hospital

Fifteen SCA patients had mild-, six had moderate- and one had severe hearing loss compared to the controls who had respectively 5 mild- and one moderate hearing loss.

Configuration of hearing loss among cases and controls attending Mulago hospital

Twenty-two SCA patients had hearing loss in at least one ear; nineteen (86%) of them had hearing loss in both ears. All patients had high frequencies of hearing loss; none had a low frequency of hearing loss.

Multivariate analysis

Participants with neural difficulty are 19.12 times more likely to experience hearing loss compared to those without neural difficulty at 95% CI: 2.4-150.3. This was statistically significant at p=0.005. Other factors described in the multivariate model above were not statistically significant.

Table 4: Multivariate model of factors independently associated with hearing loss among cases and controls.

	Cases			Controls		
Variables	OR	(95% CI)	P value	OR	(95% CI)	P value
Age (years)						
5-10	1			1		
>10	2.65	0.8-8.5	0.1	6.834	0.9-52.5	0.065
Sex						
male	2.13	0.7-6.4	0.176	1.033	0.1-7.9	0.975
female	1			1		
Coma						
No	1					
Yes	0.81	0.1-5.5	0.832	1		
Dizziness						
No	1					
Yes	2.97	0.9-10.3	0.087	1		
Neurologic motor deficit						
No	1					
Yes	19.12	2.4-150.3	0.005*	1		

DISCUSSION

The results of this study show that patients with SCD have high hearing thresholds compared to non-sickle cell patients. Prevalence of hearing loss found among children with SCD was 17% compared to 5% of the controls. These findings are in agreement with findings from previous studies which showed hearing impairment ranging between 11-41%. 4.6.7

In this study, CHL was found to be 25%, SNHL 39% and MHL 14% among the cases compared to 7% of conductive hearing loss, 7% sensorineural and 7% mixed among the controls. The prevalence of SNHL was higher, which is consistent with the finding of other studies. ¹³⁻¹⁶ However, this differs from one study done in Nigeria in 2008 by Alabi et al, in which the prevalence of SNHL was 3.8% and the CHL prevalence was 27.5%, this was due to otitis media with effusion. ¹⁷

Majority (95%) of the cases with hearing loss; 21 patients (75%) in total had hearing loss ranged from mild to moderate and one patient (4%) was found with severe hearing loss compared to 5 controls (17%) who had mild hearing loss and 1(4%) who had moderate hearing loss. This was consistent with a study conducted by Elola et al. ¹⁸ It noted hearing impairment at threshold 30 to 65 dB. However, no severe hearing loss was found in the above study. A possible neural cause of SNHL was suggested

by some experts in patients with a history of ischemic stroke.¹⁸ This could be investigated using ABR which is not yet available at Mulago National Referral Hospital (MNRH).

All patients in our study had developed high-frequency hearing loss. These findings are consistent with the studies done by Alabi et al Although the pathophysiology of the hearing loss in SCD is not yet well known, this seems to confirm the postulated theory, that SCD destroys basal hair cells where high frequencies are recorded and later the hair cells which are responsible for the recording of lower frequencies. 9,17 However, it contrasts with the findings of the study carried by Elola et al in Ivory Coast in which the low frequencies were more affected (58% of cases). Majority of the cases (86%) had bilateral hearing loss. This finding is similar to what was found in other studies. No statistically significant association in the difference of configuration was found between the cases and the controls.

The association of hearing loss with the presentation of a neurologic motor deficit (p=0.05) on examination was found statistically significant among SCD patients in the current study. This is similar to the study done by Otavio et al, whose findings showed a significantly higher prevalence of SNHL among patients with neurological sequelae compared with those who did not (p=0.2). 14

Limitations

The reliability of data concerning the history of the Sickle cell disease and the factors associated was not confirmed because it was based on self-report. The lack of equipment and limited resources could not allow the use of other objective investigations like tympanometry, ABR and OAEs in the determination of hearing loss among SCA patients for this study.

CONCLUSION

The prevalence of hearing loss is high among SCA patients in particular SNHL which is irreversible most of the time, so much effort must be made to assess their hearing early and regularly. We hope that, in the management of sickle cell patients at Mulago sickle cell clinic, regular audiometric assessment should be routinely done. This will help to identify early signs of auditory complications, then referral for rehabilitation and thus lead to early interventions.

We therefore recommend: further research to understand the pathophysiology of the hearing loss in SCA and possible prevention, regular otologic and audiological assessment of SCA patients should be done, whether the patient presents with hearing problems or not, at least once a year to enhance early detection of hearing loss, patients found with hearing loss should be rehabilitated as soon as possible and facilitated to access speech and language care, sickle cell patients who present with neurologic motor deficit are at high risk to develop hearing loss; so they should be tested early for hearing loss, and then rehabilitated effectively if already affected.

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Institutional Ethics Committee

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